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Case Report

Giant coronary artery aneurysms in a Japanese octogenarian: The oldest case of Kawasaki disease?

Stephen G. Chun (BS)[a,*], Jennifer A. Armstrong (MS)[a], Derek K. Pang (MD)[a], Jeffrey M. Lau (MD)[b], Ralph V. Shohet (MD)[c]

[a] John A. Burns School of Medicine, University of Hawaii, 651 Ilalo St., 3rd Floor, Honolulu, HI 96813, USA
[b] Department of Surgery, John A. Burns School of Medicine, University of Hawaii, Honolulu, HI, USA
[c] Center for Cardiovascular Research, Department of Medicine, John A. Burns School of Medicine, University of Hawaii, Honolulu, HI, USA

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Hawaii

Summary Kawasaki disease (KD) is a leading cause of non-atherosclerotic coronary artery aneurysms and, less commonly, peripheral artery aneurysms. We report an 81-year-old Japanese man from Hawaii with a history of an abdominal aortic aneurysm, bilateral iliac aneurysms, and an ambiguous right atrial cystic mass. The patient developed new-onset atrial fibrillation during lithotripsy. Angiography and magnetic resonance imaging revealed giant coronary artery aneurysms of the right coronary artery (RCA) and left anterior descending artery, and a thoracic aortic aneurysm. The RCA aneurysm was greater than 2 in. in diameter at the time of operation. Although we cannot confirm whether the patient had KD during childhood, this is the most likely diagnosis in the absence of a connective tissue disorder, systemic vasculitis, or atherosclerotic risk factors. This patient may represent the oldest case of KD, predating the earliest known case by more than 20 years. This case sheds light on the historical epidemiology of KD and its clinical course, especially regarding late vascular sequelae.

Background

Coronary artery aneurysms are rare, and small aneurysms are found incidentally in 1–4% of cardiac angiography [1]. Although the underlying mechanisms responsible for the development of coronary aneurysms are poorly understood, their pathogenesis is thought to be mediated by vasculitis characterized by the dysregulation of transforming growth factor-β, matrix metalloproteinases, and tumor necrosis factor-α, resulting in the disruption of vascular integrity [2]. Major causes of coronary aneurysms include atherosclerosis, connective tissue disorders, and Kawasaki disease (KD) [1].

KD, also known as mucocutaneous lymph node syndrome, typically manifests with fever, maculopapular rash, conjunctivitis, and peripheral edema in children younger than 8 years of age. It is the most common vasculitis of infancy and childhood, and the development of giant coronary

* Corresponding author. Tel.: +1 808 288 0001.
E-mail address: sgschun@hawaii.edu (S.G. Chun).

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aneurysms is the most feared and dangerous sequela of the acute syndrome. Prior to this report, the first probable cases of KD were reported in Japan during the 1950s, followed by its identification throughout the world during the 1960s and 1970s, predominantly among children of Asian descent [3]. Although the etiologic agent of KD has remained elusive, the history of the disease suggests that a hypothesized infectious entity from Japan spread throughout the world during the 1970s via Hawaii [3].

While KD is a self-limiting vasculitis, coronary aneurysms frequently developed prior to modern therapy in 20—25% of patients that clinically manifest as sudden myocardial infarctions or coronary artery rupture [3]. Despite its predilection for the coronary arteries, KD has also been reported to cause systemic artery aneurysms in a subset of patients [4—6]. Although the acute and subacute sequelae of KD are well characterized, its long-term prognosis is unknown. Moreover, recent reports indicate that systemic artery aneurysms can develop years after acute KD [6].

We report a case of a Japanese octogenarian from Hawaii with giant coronary artery aneurysms and a thoracic aortic aneurysm, with a past medical history of an abdominal aortic aneurysm and bilateral iliac aneurysms. This patient is suspected to have developed KD during the late 1920s, which would predate the earliest known cases of KD by more than 20 years.

Case report

In October of 2007, an 81-year-old Japanese man from Hawaii developed new-onset atrial fibrillation with hypotension during lithotripsy for nephrolithiasis. His medical history was significant for bilateral iliac aneurysms and an abdominal aorta aneurysm that were surgically repaired 14 years prior to presentation. An abdominal and iliac aortogram performed shortly after the surgical repair 14 years previously demonstrated patent renal and femoral arteries without aneurysmal changes.

Two years prior to the current presentation, echocardiography identified an ambiguous cystic mass located in the body of the right atrium during workup for an asymptomatic ill defined murmur (Fig. 1). The echocardiogram was otherwise unremarkable and thallium myocardial scanning showed no areas of ischemia at that time. In the absence of symptoms, further evaluation was not pursued in this elderly gentleman.

The patient had none of the common risk factors for atherosclerosis such as hypertension, hypercholesterolemia,

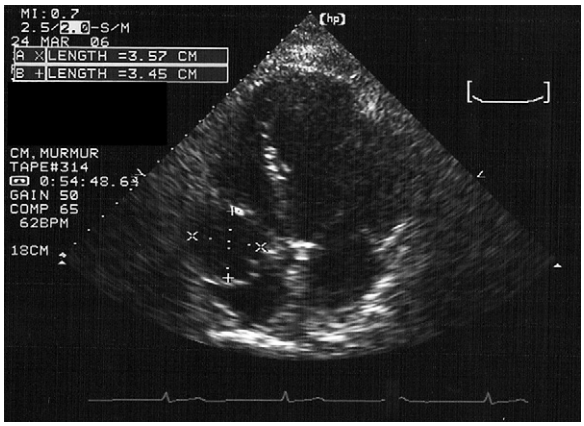

Figure 1 Echocardiography apical four-chamber view showing a spherical mass (3.6 cm × 3.5 cm) adjacent to the right atrium.

history of smoking, or a family history of coronary artery disease. The patient was normotensive at several recent clinic visits, and his most recent lipid profile included a total cholesterol of 182 mg/dL, triglycerides 59 mg/dL, high density lipoprotein cholesterol 79 mg/dL, and low density lipoprotein cholesterol 91 mg/dL.

Physical exam at this most recent presentation did not reveal signs of a connective tissue disorder or systemic vasculitis. The patient had a normal limb-to-body ratio and no joint laxity or lens dislocation suggestive of Marfan's or Ehlers-Danlos Syndrome. He also lacked mucosal ulceration, uveitis, or visual changes suggestive of Bechet's Disease. Laboratory evaluation included a C-reactive-protein (CRP) level of 2.0 mg/L (normal 0—10 mg/L), erythrocyte sedimentation rate (ESR) of 11 mm/h (normal 0—20 mm/h), and negative Venereal Disease Research Laboratory test for syphilis. In the presence of normal white blood cell count, ESR, and CRP, and absence of history or physical exam findings suggestive of a pro-inflammatory state, further laboratory tests were not performed for collagen vascular diseases.

Coronary angiography revealed giant aneurysms of the proximal right coronary artery (RCA) and left anterior descending (LAD) arteries, and an aneurysm of the ascending thoracic aorta (Fig. 2). Angiography also revealed stenoses of the RCA and LAD prior to the aneurysms and complete obstruction of blood flow distal to the giant RCA aneurysm. The first diagonal artery was diffusely diseased. Magnetic resonance imaging (MRI) confirmed the presence of a giant

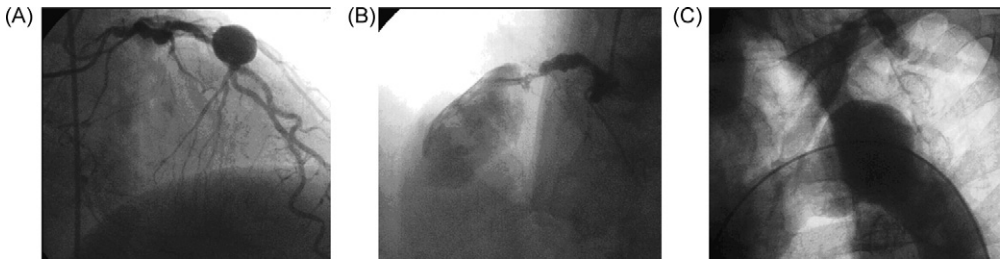

Figure 2 (A) Aneurysm of the left anterior descending artery. (B) Giant aneurysm of the right coronary artery. (C) Aneurysm of the thoracic aorta.